Interventional Management of Lymphatic Morbidity in Patients With CHD

Maxim Itkin MD, FSIR
Professor of Radiology and Pediatrics
Hospital of University of Pennsylvania
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Lymphatic System Function

Removal of the transudate (proteins/water) from the tissues back into systemic circulation
Lymphatic System-Flows

Lymphatic Systems

- Liver
- Intestine
- Soft tissue
Lymphatic Consequences of Congenital Heart Surgery

- Hepatic Venous Pressure
- Liver Lymphatic Flow
- Central Lymphatic System Flow
- Lymphatic Complications
Pressure in the sinusoid is low
Need to maintain positive influx of ultrafiltrate
Hepatic sinusoids are highly permeable to albumin
It seems probable that the obstruction to flow of lymph from the thoracic duct into the blood as well as the distention of the duct from the largely increased lymph flow from the liver which is present in uncompensated heart disease may contribute to the production of edema in the rest of the body (and) to the production of hydrothorax.
Ernest H. Starling, The Arris and Bale lectures on some points in the pathology of heart disease., In The Lancet, Volume 149, Issue 3835, 1897, Pages 569-572
Size of the Thoracic Duct in Heart Failure

Congestive Heart Failure on MRL

Lymphatic Complications of Congenital Heart Surgery

- Abnormal Pulmonary Lymphatic Flow
  - Postsurgical Chylothorax
  - Plastic Bronchitis
- Abnormal Hepatoduodenal Lymphatic Flow
  - Protein Losing Enteropathy
Lymphatic Complications of Congenital Heart Surgery

There is lack of correlation between the severity of heart failure and incidence of lymphatic complications
Lymphatic Complications of Congenital Heart Surgery

There is lack of correlation between the severity of heart failure and incidence of lymphatic complications.

Increased lymphatic flow

Clinical presentation
There is lack of correlation between the severity of heart failure and incidence of lymphatic complications
Abnormal Pulmonary Lymphatic Perfusion

Congenital Lymphatic Variant

Plastic Bronchitis

Post Cardiac Surgery Chylothorax
Abnormal Pulmonary Lymphatic Perfusion

**Congenital Lymphatic Variant**

- Plastic Bronchitis

**Post Cardiac Surgery Chylothorax**
Plastic Bronchitis

- Formation of large gelatinous or rigid branching airway casts
Lymphangiography
TD Injection with Methylene Blue
Treatment - TD Embolization

Intranodal Lymphangiography
TD catheterization
TD embolization
Thoracic Duct Access
Thoracic Duct Access
TD Injection
Embolization

Microcoils

• Nester 0.018

Liquid embolic agent

• Glue - n-Butyl Cyanoacrylate (n-BCA)
Glue injection
PB Lymphatic Embolization-Outcome

- 18 Patients with “cardiac” PB
- 16 demonstrated pulmonary lymphatic perfusion
- 15/16(94%) — significant improvement of their symptoms
- Median follow-up 315 days
- One major complication-TIA
  - Lymphatic to pulmonary vein

Y Dori, M Keller, JJ Rome, MJ Gillespie, AC Glatz, K Dodds, DJ Goldberg, S Goldfarb, J Rychik, M Itkin
Post Pediatric Cardiac Surgery Chylothorax

Retrospective review of 25 patients

MR lymphangiography and Intranodal Lymphangiography

Three groups of patients

- Abnormal Pulmonary Lymphatic Perfusion: 14 patients
- Central Lymphatic Flow Disorder: 9 patients
- Traumatic Chylous Leaks: 2 patients

Pulmonary Lymphatic Perfusion Syndrome

Central Lymphatic Flow Disorder

Absent/Diminutive TD
Combination of ascites and pleural effusion
Tissue edema
Dermal collaterals
Under 1 year of age
4/9 patient had TD ligation

Central Lymphatic Flow Disorder

Results

PLPS and Trauma - 16 patients

- TD embolization-all patients
- All 16 patients had resolution of chylothorax after embolization

Central Lymphatic Flow Disorder - 9 patients

- TD embolization- 6 patients
  - Clinically unsuccessfully
- One patient underwent lympho-venous anastomosis
  - Resolution of chylothorax
- 7 patients deceased

Central Lymphatic Flow Disorder

TD-Venous Anastomosis
TD-Venous Anastomosis
TD-Venous Anastomosis
Liver Lymphatic System - Anatomy

Liver lymph 40% of the flow in TD

High concentration of the albumin

Flow increases significantly in heart failure
Communication of the Liver Lymphatics with TD
Protein Losing Enteropathy

- Severe loss of serum proteins into the intestine
PLE Pathophysiology Concept

Physiology:

- Liver generates albumin and delivers it into the blood stream through lymphatic system
- Liver lymph has a high concentration of proteins
- The lymphatic flow in the liver increases significantly in patients with CHF

Hypothesis:

- The loss of the proteins in PLE happens from the liver lymph leaking into the intestine
PLE Treatment Concept

Perform liver lymphangiogram

• Leak?

If leak attempt to embolize
Liver Lymphangiogram PLE

Liver Lymphangiogram PLE

PLE Embolization Outcome

- 8 patients
- 3 patients temporary response
- 3 patients sustained response
- 363 days (range 84-1005)
- 2 duodenal bleeding

Embolization of Hepatoduodenal Communications
Conclusions

Lymphatic imaging is crucial in understanding of pathophysiology.

Understanding of the lymphatic variants is essential to explain some of the symptoms in patients with congenital heart disease.

Improving the outcomes.

Preventions and predictions.